TEXAS CHILDREN'S HOSPITAL
EVIDENCE-BASED OUTCOMES CENTER
Acute Management of Congenital Diaphragmatic Hernia
Evidence-Based Guideline

Definition: Congenital diaphragmatic hernia (CDH) is a relatively common malformation in which there is failure of complete fusion of the diaphragm during the prenatal period. This leads to protrusion of abdominal organs into the thoracic cavity which can result in lung hypoplasia and respiratory failure at birth. (1) CDH manifests on the left side in 75% of cases, which leads to possible herniation of the small and large bowel, the spleen, the stomach, the left lobe of the liver and, rarely, the kidney. Right-sided CDH can include herniation of the right lobe of the liver and possibly the bowel and/or kidney. (2) A bilateral defect is seen rarely. (3) Severity of disease depends upon the degree of abdominal organ herniation and lung hypoplasia.

Epidemiology: CDH occurs in approximately 1 in 3000 live births. Modern advances in technology and clinical practice for this condition has improved the reported overall survival to 79% (range: 69 – 93%), with isolated CDH having the highest survival rates.(4) In approximately 40% of cases, other associated anomalies are present in CDH patients, (3) including cardiac anomalies that are present in 25% of cases. (5) Musculoskeletal, neural tube, abdominal wall, craniofacial defects and urinary tract anomalies have been found in CDH patients.

Etiology: A variety of genetic abnormalities have been shown to cause isolated and non-isolated CDH. These abnormalities include large chromosome anomalies, microdeletions, microduplications, and mutations affecting single genes. CDH has also been shown to be a feature in a variety of genetic syndromes.

Inclusion Criteria:
- Neonate diagnosed with CDH

Exclusion Criteria:
- Infant not diagnosed with CDH

Differential Diagnosis: Chest x-ray results of CDH patients may be similar to that of a congenital cystic adenomatoid malformation (CCAM). The identification of abdominal organs in the thoracic cavity and a paucity of bowel in the abdomen solidifies diagnosis of CDH. (2)

Diagnostic Evaluation: The majority of CDH cases are diagnosed during antenatal ultrasound procedures during the second or third trimester. (6) Once the diagnosis is made, radiological ultrasound (US) and fetal magnetic resonance imaging (MRI) allows for determination of severity of disease by determining the percentage of herniated liver (%LH), observed-to-expected fetal lung volumes (O/E-TFLV) and the lung-head ratio (LHR). After birth, the diagnosis of CDH is made based upon signs and symptoms with confirmation by x-ray. (2)

A detailed history and physical examination should be completed to assess the severity of the disease, to identify other congenital anomalies and to identify features that could suggest a specific genetic syndrome.

History: Assess for
- Relevant maternal history
- Results of antenatal imaging studies
- Results of genetic tests

Physical Examination: The following physical examination findings may be present in infants with congenital diaphragmatic hernia. Assess for: (3)
- Scaphoid abdomen
- Absence of breath sounds on the ipsilateral side
- Barrel-shaped chest
- Shifted cardiac sounds
- Bowel sounds in the chest

Laboratory Tests:

Lab	Delivery Room	Immediately upon NICU Admission	At 24 Hours of Life
Glucose	X		
Blood culture*		X	
Arterial Blood Gas#		X	
Serum Lactate#		X	
CBC with Plat and Diff		X	
Type and Screen		X	
BNP			X†
Chem 7			X
Bilirubin Panel			X
Newborn Screen			X
Chromosomal Microarray Analysis			X

*If concerns for sepsis
ABG & Lactate to be ordered at Q1 to Q4 (or greater) intervals depending on clinical status in first few hours of life
† BNP should be drawn within the first 24 hours of life and before surgery

Ongoing Laboratory Tests
- B-type natriuretic peptide (BNP) testing should be measured on day of life 1, postoperative day 1 and weekly thereafter. More frequent monitoring of BNP levels may be obtained as clinically indicated. (7-13)

Imaging Studies:

Study	Timing
Chest X-ray including abdomen	STAT upon admission to NICU
Head Ultrasound	Within 24 hours of admission unless otherwise indicated; Upon arrival for high risk patients (see Appendix)
Echocardiogram	Within 24 – 48 hours of admission unless otherwise indicated; Upon arrival for high risk patients (see Appendix)

Prenatal Assessments

- Depending on the gestational age at the time of CDH diagnosis, it is recommended that a fetal echo be performed within a week or two after the diagnosis is made to rule out structural heart disease, especially if Fetal tracheal occlusion (FETO) is being considered.
- If FETO is not being considered, a fetal echo may be performed at a convenient time, preferably between 24-32 weeks gestation.
- The percentage of herniated liver (%LH) and observed-to-expected fetal lung volumes (O/E-TFLV) obtained from MRI should be used to stratify CDH severity which can be used in prenatal counseling to educate families on disease severity and mortality risks. (14-26)
- Practitioners should measure and document %LH, O/E-TFLV, the side of the defect, total lung volume (TLV), lung-to-head ratio (LHR), observed-to-expected lung-to-head ratio (O/E-LHR), stomach position and lung-to-thorax ratio for risk stratification, research, benchmarking and outcomes reporting in CDH patients. (14-37)
- There should be a case-by-case determination of the management plan for CDH patients with comorbidities identified prenatally. This should be preferably discussed with the family prior to delivery and involve P-PACT if necessary. Limitations in care should be discussed in the antenatal visit.

Postnatal Echo

- It is recommended that a post-natal echocardiogram be performed within the first 24-48 hours after birth. It may be obtained earlier if the neonate exhibits a sub-optimal response to the usual therapy, especially if a pre-natal echo was sub-optimal or not performed. (38-42)
- A post-natal echo may be obtained electively, at a later, convenient time, if the neonate does not exhibit undue difficulties in ventilation and oxygenation, especially if a good quality fetal echo was performed.

Cardiac Cath – Acute Phase

- Cardiac cath is not necessary if left-to-right atrial and ductal flow is noted by echocardiography, as this would suggest adequate and favorable response to therapy.
- In the neonatal period, cardiac cath may be considered upon the recommendation of a cardiologist, if the clinical course is suggestive of pulmonary venous abnormalities and other less invasive imaging modalities are not appropriate. (38-42)
- Cardiac cath may be necessary if there is a need to perform a transcatheter cardiac intervention (i.e. – the need to stent a PDA in the presence of Coarctation) in a neonate deemed not to be a surgical candidate.

Cardiac Cath – Chronic Phase with Persistent Pulmonary Hypertension

- Cardiac cath should not be performed in a patient who demonstrates normal RV function and left to right atrial and ductal flow by echocardiography, as this would suggest adequate and favorable response to therapy.
- A cardiac cath may be considered, especially in the presence of RV dysfunction (cor pulmonale) and predominant right to left atrial and ductal level shunting, if the response to pulmonary vasodilator therapy is sub-optimal and other pharmacologic options are being considered. (38-42)
- In the presence of severe pulmonary hypertension and RV failure, cardiac cath is recommended if an intervention such as an atrial septostomy or ductal stenting is being considered. Consider cardiac cath if there is left ventricle dysfunction present to better characterize the anatomy.
- A cardiac cath may be necessary to better delineate the anatomy when the patient has other anatomical issues that are not able to be evaluated by echo.

Other Imaging Tests

- Pulmonary venous abnormalities may be difficult to rule out by fetal or post-natal echocardiography in a patient with CDH. Additional imaging modalities, such as a contrast chest CT, cardiac MRI or angiography by cardiac cath may be recommended by the clinician if echocardiography is unable to definitively rule out abnormalities of pulmonary venous return. This is especially true in the profoundly hypoxemic neonate who does not respond to the usual measures, and in the presence of pure right to left shunting in the atrial or ductal level.
- A small percentage of the CDH population may require additional imaging studies such as Ventilation Perfusion (V/Q) scan.

Critical Points of Evidence

Evidence Supports

Prenatal Testing

- The percentage of herniated liver (%LH) and observed-to-expected fetal lung volumes (O/E-TFLV) obtained from MRI should be used to stratify CDH severity which can be used in prenatal counseling to educate families on disease severity and mortality risks in CDH patients. – Strong recommendation, moderate quality evidence (14-26)
- Practitioners should measure and document the %LH, O/E-TFLV, side of the defect, total lung volume (TLV), lung-to-head ratio (LHR), observed-to-expected lung-to-head ratio (O/E-LHR), stomach position and lung-to-thorax ratio for risk stratification, research, benchmarking and outcomes reporting in CDH patients. – Strong recommendation, moderate quality evidence (14-37)
- A pre-established objective criteria should be used to score the severity of cardiac lesions. The presence of a major cardiac anomaly is associated with an increased risk of mortality. – Strong recommendation, low quality evidence (32,43-45)
- The oxygenation index should continue to be used for management decisions in CDH patients. – Strong recommendation, low quality evidence (46-49)

- The Brindle Prediction Score (see appendix) should be used to educate families on the categorical (low, intermediate or high) mortality risk in CDH patients. – Strong recommendation, low quality evidence [14,50]

Initial Management

- Clinicians may consider the use of surfactant in CDH patients with a history of Fetal Endotracheal Occlusion (FETO) or preterm birth (< 37 weeks gestation). – Weak recommendation, low quality evidence [5,51-57]
  - Remarks: The fetus that undergoes FETO can have a decrease in type II pneumocytes which can cause surfactant deficiency. Patients receiving this procedure during the prenatal period may benefit from postnatal surfactant administration. Response to therapy guides dosing of surfactant. Unless there were negative consequences to initial administration, clinicians should consider a second dose. If the patient does not exhibit a positive response to the first two doses of surfactant, do not administer additional doses. [58,59]

Laboratory Testing and Imaging

- B-type natriuretic peptide (BNP) testing should be measured on day of life 1, postoperative day 1 and weekly thereafter. More frequent monitoring of BNP levels may be obtained as clinically indicated. – Strong recommendation, very low quality evidence [7-13]
- Echocardiogram should be performed within 24 to 48 hours of life, one week after CDH repair, and within four weeks of discharge. Additional follow-up echos should be obtained every 1 to 2 weeks for patients with severe pulmonary hypertension until there is evidence of resolution with pulmonary pressures <1/2 systemic. – Strong recommendation, very low quality evidence [38-42]

Systemic Hypotension Management

- Titrate dopamine to manage systemic hypotension in CDH patients during the acute phase. – Strong recommendation, low quality evidence [55,60-67]

ECMO

- Administer a one-time dose of cefazolin within one hour of cannulation for ECMO in CDH patients. Do not routinely use continued prophylactic antibiotics for patients on ECMO support. – Strong recommendation with very low quality evidence [68-70]

Surgical Repair

- In non-repaired CDH patients on ECMO, surgical repair of the diaphragm should be completed within 48 hours of ECMO cannulation. – Strong recommendation, low quality evidence [1,5,55,63,64,66,67,71-78]
- In CDH patients not requiring ECMO, surgical repair of the diaphragm should be performed when the patient is physiologically stable defined as mean blood pressure normal for gestational age, preductal oxygen saturations between 85-95% on FiO2 below 50%, lactate below 3 mmol/L, urine output greater than 2 mL/kg and less than a 10% gradient between pre- and post-ductal saturations. – Strong recommendation, low quality evidence [1,5,55,63,64,66,67,71-78]
- In CDH patients that are not able to achieve a tension-free primary closure of the diaphragm, a patch repair should be completed with an appropriately sized domed patch. – Strong recommendation, low quality evidence [76-79]
- CDH patients with large defects should be repaired using an open surgical technique. – Strong recommendation, moderate quality evidence [78,82-86]
- The use of minimally invasive surgery (MIS) may be considered in patients that meet the following criteria: mean blood pressure normal for gestational age without use of inotropes, pre-ductal oxygen saturations between 85 – 95% on FiO2 below 40% without use of inhaled nitric oxide, lactate below 3 mmol/L, urine output greater than 2 mL/kg/hr, and less than a 5% gradient between pre- and post-ductal saturations. – Weak recommendation, moderate quality evidence [1,5,55,63,64,66,67,71-78]

Evidence Lacking/Inconclusive

Management Plan

- Determine management plan for severe cardiac lesions first before proceeding to establish plan of care for congenital diaphragmatic hernia. – Consensus recommendation

Ventilation Strategy

- Patients with CDH should be initially ventilated with the conventional ventilator using the AC/VG mode with initial settings of PEEP 5-6 cmH_2O, TV 4 – 5 mL/kg, back-up rate 40 breaths/min, IT 0.3 seconds, and FiO2 adjusted for target preductal saturations of ≥80%. – Strong recommendation, low quality evidence [5,55,63-67,78,87-91]
- Tidal volume should be adjusted to meet optimal physiological monitoring parameters. – Strong recommendation, low quality evidence [5,55,63-67,78,87-91]
- Clinicians should consider switching the mode of ventilation to HFOV for CDH patients that cannot achieve target PCO_2 on conventional ventilation with PIP ≤ 28. – Weak recommendation, low quality evidence [5,55,63-67,78,87-91]
  - Once on HFOV, increase MAP (to a max MAP of 17) and DeltaP as required to achieve physiological monitoring parameters.
- Clinicians should consider ECMO for CDH patients that cannot achieve saturations and/or blood gas targets with maximal HFOV support. – Weak recommendation, low quality evidence [5,55,63-67,78,87-91]
- Patients with CDH should have the following targets for physiological monitoring parameters: pre-ductal oxygen saturations ≥80% for the first two hours of life. Thereafter, pre-ductal saturations should be ≥85%; pH > 7.20; pCO_2 50 to 70; and pO_2 40 to 90. – Strong recommendation, very low quality evidence [5,55,63-67,78,87-91]
- Patients with CDH should be initially ventilated with 100% FiO_2. – Consensus recommendation
- Pre-ductal saturations should be targeted at ≥70% for the first ten minutes after birth; increasing to ≥80% for the first two hours of life. Thereafter, pre-ductal oxygen saturations should be ≥85%. – Consensus recommendation

Venous Access and Fluid Management

- Peripheral venous access should be established in the delivery room for CDH patients. – Consensus recommendation
- Umbilical lines and elective/non-emergent procedures should be completed in the NICU. – Consensus recommendation
- An umbilical venous catheter (UVC) and an appropriately placed umbilical arterial catheter (UAC) should be initially inserted in CDH patients with a need for central venous access. If correct UVC position cannot be achieved, a temporary low-lying UVC can be initially utilized until a sufficient alternative is available. - Strong recommendation, low quality evidence (102-103)
- A peripherally inserted central catheter (PICC) and an arterial line should be placed by the NICU Vascular Access Team (VAT) in CDH patients with a need for long-term central venous access once stabilization occurs. – Consensus recommendation
- Initial fluid intake for CDH patients should be 65 mL/kg/day. – Consensus recommendation

Pulmonary Hypertension Management

- In infants with congenital diaphragmatic hernia with an oxygenation index (OI) >25, preductal saturations <90% on 100% FiO_2 or a gradient between pre- and postductal saturations ≥ 10%, consider the use of inhaled nitric oxide to improve oxygenation. – Weak recommendation, low quality evidence (5,38,55,63-67,78,104-109)
- In infants with congenital diaphragmatic hernia, consider the use of sildenafil for continued clinically significant pulmonary hypertension beyond the acute phase as evidenced by a preductal/postductal saturation difference of > 10 points, in those infants who fail weaning from inhaled nitric oxide or to facilitate weaning from nitric oxide. – Weak recommendation, low quality evidence (5,38,55,63-67,78,110,111)
- In infants with congenital diaphragmatic hernia requiring VA ECMO, inhaled nitric oxide should be discontinued once VA ECMO support is initiated and restarted when recruiting the lungs in preparation for discontinuing ECMO. – Strong recommendation, very low quality evidence (55, 112-114)
- In patients on VV ECMO, consider the use of inhaled nitric oxide throughout ECMO run. – Weak recommendation, very low quality evidence (55, 112-114)
    - Remarks: Response to inhaled nitric oxide should be assessed in all users within one hour of initiation. Responders will exhibit a >5% increase in preductal saturations, an increase in PaO_2 by 10 torr (obtained from the same pre- or post-ductal source) or a decrease in pre/post ductal saturation gradient to <10%. Inhaled nitric oxide should be weaned based upon patient classification as responder or non-responder.

Systemic Hypotension Management

- Consider administration of hydrocortisone (1mg/kg IV every 8 hours) in CDH patients on a dose of greater than or equal to 10 mcg/kg/min of dopamine with continued systemic hypotension. – Weak recommendation, very low quality evidence (55,63-67,115-117)
- Consider the use of epinephrine in CDH patients on maximum dopamine dose and hydrocortisone. – Weak recommendation, very low quality evidence (55,63-67,118)
    - Remarks: If the patient has vasopressor refractory hypotension, an echo should be obtained and the treated according to findings.

Evidence Against

Prenatal Testing

- The Brindle Prediction Score (see appendix) should not be used for individual patient decisions regarding the use of ECMO or other medical management. – Strong recommendation, low quality evidence (14,50)
- The SNAP II Score, Wilford Hall/Santa Rosa Clinical Prediction Formula, Congenital Diaphragmatic Hernia Study Group (CDHSG) formula, the best $PaCO_2$ at 1 hour of life and 24 hours of life, and the highest preductal O_2 saturation value during the first 24 hours of life should not be used to predict survival or make decisions about treatment options for CDH patients. – Strong recommendation, low quality evidence (46,119-130)

Initial Management

- Surfactant should not be routinely used in the non-FETO term CDH patient at birth. – Strong recommendation, low quality evidence (5,51-57)
- Information obtained from near infrared spectroscopy (NIRS) readings should not be used for patient care management. – Strong recommendation, low quality evidence (131)

Pulmonary Hypertension Management

- Prostaglandin E (PGE) should not be routinely used in the acute phase of CDH treatment. – Strong recommendation, very low quality evidence (5,38,55,63-67,78,132,133)
- Epoprostenol should not be routinely used for the treatment of pulmonary hypertension in neonates with congenital diaphragmatic hernia. - Strong recommendation with very low quality evidence (5,38,55,63-67,78,134-136)

Imaging

- Cardiac cath is not recommended in the acutely presenting CDH neonate in whom congenital heart disease has been ruled out by echocardiography. – Strong recommendation, very low quality evidence (38-42)

Ventilation Strategy

- Heliox should not be routinely used in CDH patients. – Strong recommendation, low quality evidence (137-141)

Condition-Specific Elements of Clinical Management

Initial Management in the Delivery Room:

- Intubate immediately and ventilate with conventional ventilator on AC/VG mode on 100% FiO2 (1,5,55,63,64,66,67,71-78)
    - PEEP: 5 – 6 cmH_2O
    - TV: 4 – 5 cc/kg
    - MAX PIP: 28
    - Back-up rate: 40 breaths/minute
    - IT: 0.3 seconds
    - FiO2 – adjust for target pre-ductal saturations of ≥80%
- Insert 10 french (FR) repogle and place to low-intermittent suction.
- Establish peripheral venous access and begin 10% dextrose solution. Total initial total fluid intake should be 65 mL/kg/day.
- Continually assess for respiratory distress or low pre-ductal saturations. Pre-ductal saturations should be targeted at ≥70% for the first ten minutes after birth; increasing to ≥80% for the first two hours of life.
- Adjust TV to improve distress and increase pre-ductal saturations to optimal targets
- If continued distress, adjust TV and PEEP.
- Surfactant should not be routinely used. If history of FETO or <37 weeks gestation, consider surfactant administration.(5,51-57)
- If distress continues, adjust ventilator and/or hand bagging as needed. Plan for HFOV following admission to WT NICU.
- Umbilical lines and elective procedures should be completed in the NICU.
- Goal for timespan of initial resuscitation in L&D is 30 minutes or less.

Acute Phase (Birth to Surgical Repair):

General

- Admit to NICU 4
- 10 FR repogle to low-intermittent suction (LIS) if not already done
- Insert appropriately placed UVC and UAC. If umbilical lines not appropriately placed, obtain a PICC line and/or PAL. (102-103)
- Consult ECMO surgeon, Neo ECMO clinician, ECLS Primer and Pulmonary Hypertension team.
- Monitor pre- and post-ductal saturations
- Administer erythromycin and Vitamin K

Ventilation Management (5,55,63-67,78,87-91)

- Tidal volume should be adjusted to meet optimal physiologic parameters of:
    - pre-ductal saturations ≥85% after two hours of life
    - pH >7.20
    - pCO2 between 50 – 70
    - pO2 between 40 – 90
- Consider switching mode of ventilation to HFOV if patient cannot achieve optimal physiologic parameters on PIP ≤28.
- Initial HFOV settings are:
    - MAP 13 (or 2 above that on conventional ventilator)
    - IT 0.3
    - Hz 10
    - DeltaP sufficient to produce perceptible “jiggle” to upper adnominal area.
- Once on HFOV, increase MAP (to a max MAP of 17) and DeltaP as required to achieve physiological monitoring parameters.

Treatment for Pulmonary Hypertension

- Consider the use of inhaled nitric oxide to improve oxygenation if oxygenation index (OI) >25, preductal saturations <90% on 100% FiO_2 or a gradient between pre- and postductal saturations ≥ 10%. (5,38,55,63-67,78,104-109)
- Response to inhaled nitric oxide should be assessed in all users. Responders will exhibit a >5% increase in preductal saturations, an increase in PaO_2 by 10 torr or a decrease in pre/post ductal saturation gradient to <10%. Inhaled nitric oxide should be weaned based upon patient classification as responder or non-responder.

ECMO

- Indications for ECMO include oxygenation index (OI) >40 on two separate measurements, PO2 persistently <40 mmHg or lactate rising above 3.
- Administer a one-time dose of cefazolin within one hour of cannulation for ECMO in CDH patients. Do not routinely use continued prophylactic antibiotics for patients on ECMO support. (68-70)

Treatment for Systemic Hypotension

- Most CDH patients should have mean BP normal for gestational age.
- Titrate continuous dopamine to achieve optimal blood pressure. (55,60-67)
- Once dopamine reaches 10 mcg/kg/min, consider administration of hydrocortisone. (55,63-67,115-117)
- Dopamine can continue to be titrated until a MAX of 20 mcg/kg/min to obtain optimal blood pressure.
- If optimal blood pressure is not obtained on MAX dopamine and hydrocortisone, consider starting continuous epinephrine. (55,63-67,118)

Surgical Repair

- Surgical repair of the diaphragm should be performed when the patient is physiologically stable defined as below. (1,5,55,63,64,66,67,71-78)
    - Mean blood pressure normal for gestational age
    - Pre-ductal oxygen saturations between 85 – 95% on FiO2 below 50%
    - Lactate below 3 mmol/L
    - Urine output greater than 2 mL/kg/hr
    - Less than a 10% gradient between pre- and post-ductal saturations
- Patients who meet the additional requirements of pre-ductal oxygen saturations between 85-95% on FiO2 40% without the use of nitric oxide, mean BP normal for age without use of inotropes and less than a 5% gradient between pre- and postductal saturations may also be considered for minimally invasive surgery. (1,5,55,63,64,66,67,71-78)
- In non-repaired CDH patients on ECMO, surgical repair of the diaphragm should be completed less than 48 hours after ECMO cannulation. (1,5,55,63,64,66,67,71-78)
- CDH patients with large defects should be repaired using an open surgical technique. (78,82-86)
- A patch repair with an appropriately sized domed patch should be undertaken if a tension-free primary closure is not achievable during surgery. (78-81)

Post-Acute / Chronic Phase:

Treatment for Pulmonary Hypertension

- Consider the use of sildenafil for continued clinically significant pulmonary hypertension beyond the acute phase as evidenced by a preductal/postductal saturation difference of > 10 points or in those infants who fail weaning from inhaled nitric oxide. (5,38,55,63-67,78,110,111)
- Consider the use of bosentan as third line treatment for refractory pulmonary hypertension in CDH patients without liver dysfunction and with the ability to receive enteral medications. (142)
- In infants with congenital diaphragmatic hernia requiring VA ECMO, inhaled nitric oxide should be discontinued after repair when the patient is on ventilator rest settings and restarted when recruiting the lungs in preparation for discontinuing ECMO. Inhaled nitric oxide should be weaned based upon patient classification as responder or non-responder. (55,112-114)

Discharge Criteria:

Discharge planning should begin upon admission and continue throughout hospital stay. The decision to discharge should be discussed with the pulmonary and surgical teams. Coordination of discharge is essential as some treatments may take as long as four weeks for approval from payers.

- Stable cardio/respiratory for at least two weeks that can be safely delivered in the home environment.
  - FiO_2 ≤40% with home ventilator
  - NC 1 liter per minute (LPM) or less
- Stable nutrition program with documented weight gain and growth velocity.
- Appropriate training and documented education of family or guardian.
- Prescriptions should be filled and in the possession of parent or guardian at the time of discharge.
- Designated primary care physician and follow-up plan with appropriate teams.
- Consider rooming-in for patients with complicated home care

Consults/Referrals:

- Surgery
- Pulmonary
- Cardiology
- Pulmonary Hypertension Team
- Genetics as indicated

Follow-Up Care:

- Primary Care Physician
- Surgery
- Pulmonary Hypertension Team
- Developmental

Outcome Measures:

Process-

- Time of birth to admit to NICU
- Rate of antenatal consults by neonatology or relevant service (Cardiology, Surgery)
- Rate of surfactant use in the first 72 hours of life
- Timing of first echo

Outcome-

- Mortality rate
- Mean and median duration of ventilation
- ECMO rate
- Length of stay
- Duration of ECMO
- ECMO complications
- Rate of discharge on pulmonary hypertension treatment
- Rate of discharge on enteral feeding

Medication Dosing

Surfactant [143]

Surfactant should not be routinely used in the non-FETO term CDH patient at birth. Clinicians may consider surfactant in CDH patients with a history of Fetal Endotracheal Occlusion (FETO) or preterm birth (<37 weeks gestation).	
Curosurf Inhalation	**Route: Inhalation** 2.5 mL/kg via endotracheal tube for the first dose. Decrease dose to 1.25 mL/kg for subsequent doses. Response to therapy guides dosing of surfactant.

Systemic Hypotension Treatment [143]

DOPamine	**Route: Continuous IV infusion** 5 mcg/kg/min Titrate to MAX of 20 mcg/kg/min
Hydrocortisone Note: Consider administration of hydrocortisone once dopamine is titrated to 10 mcg/kg/min or greater.	**Route: IV** 1 mg/kg every 8 hours
EPINEPHrine	**Route: Continuous IV infusion** 0.05 mcg/kg/min Titrate to MAX of 1 mcg/kg/min

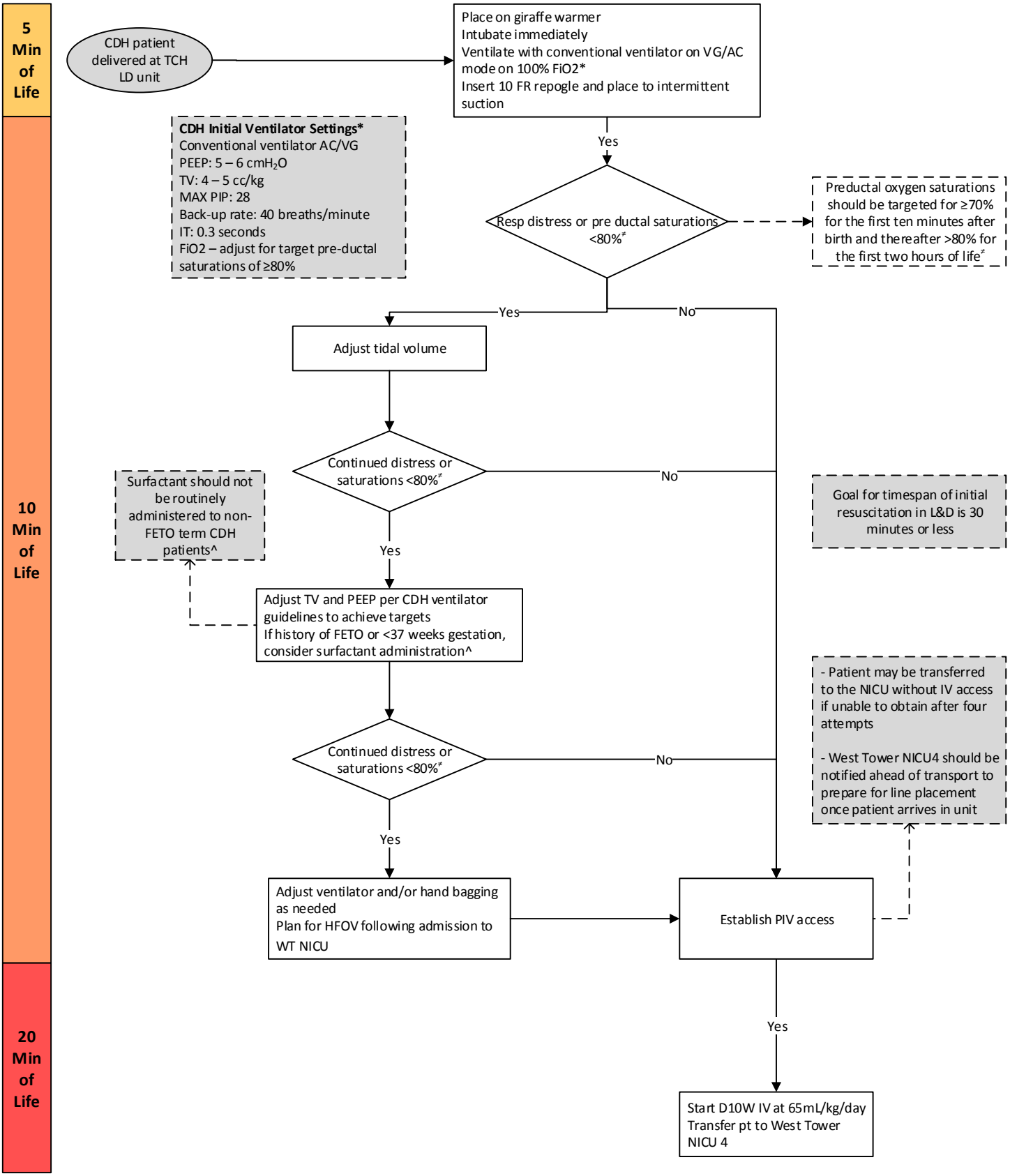
Pulmonary Hypertension Treatment – Acute Phase [143]

Nitric Oxide	**Route: Inhalation** Start with initial dose of 20ppm; titrate down to lowest effective dose

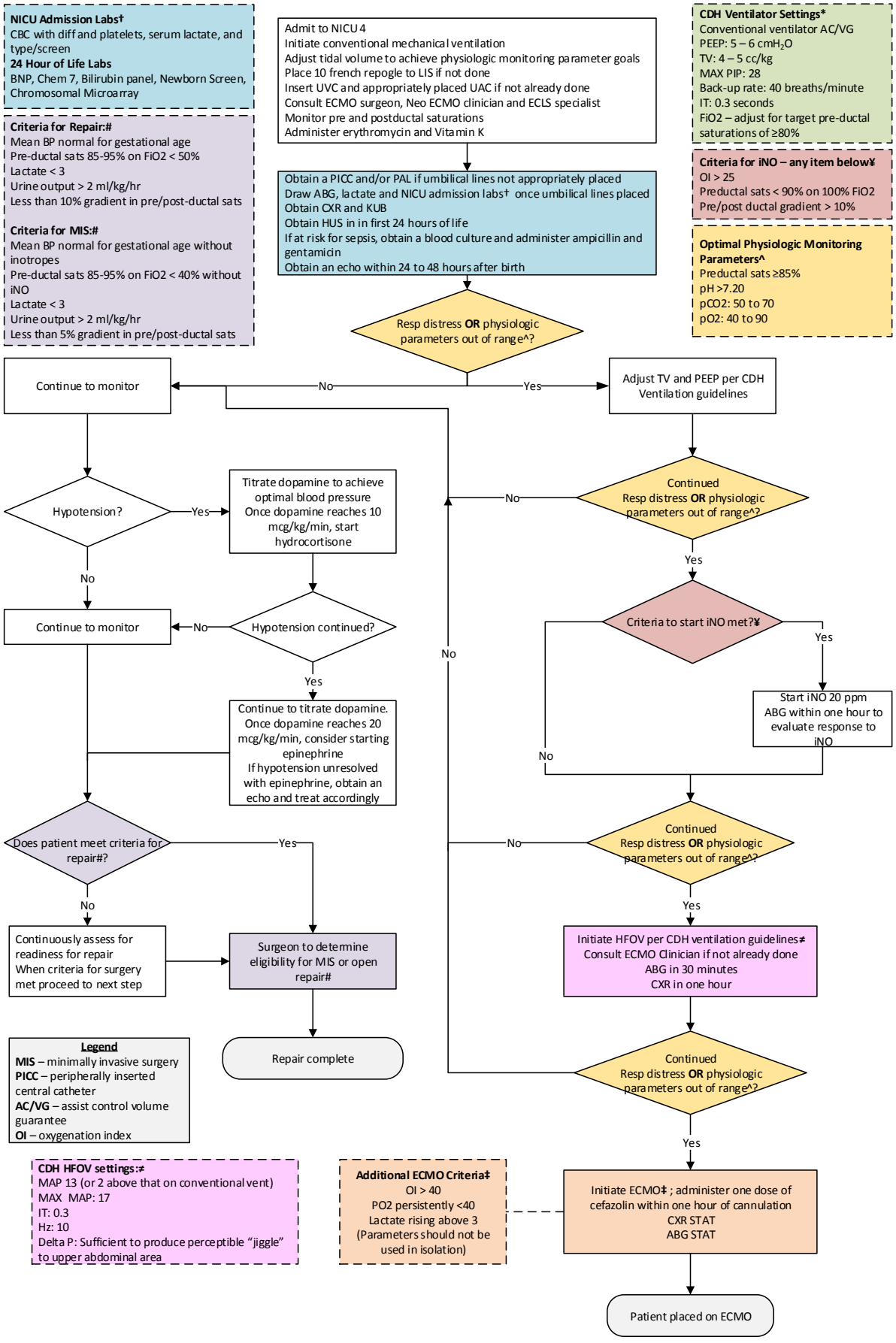
ECMO Preparation [143]

CeFAZolin	**Route: IV** 25 - 30 mg/kg IV ONCE 30 – 60 minutes before procedure

TCH Evidence-Based Outcomes Center
Clinical Algorithm for Initial Management of Congenital Diaphragmatic Hernia Patients
Labor and Delivery Algorithm



TCH Evidence-Based Outcomes Center
Clinical Algorithm for the Acute Management of Congenital Diaphragmatic Hernia Patients Prior to ECMO
Inpatient Algorithm



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Clinical Standards Preparation

This clinical standard was prepared by the Evidence-Based Outcomes Center (EBOC) team in collaboration with content experts at Texas Children's Hospital. Development of this clinical standard supports the TCH Quality and Patient Safety Program initiative to promote clinical standards and outcomes that build a culture of quality and safety within the organization.

Congenital Diaphragmatic Hernia Content Expert Team
Caraciolo Fernandes, MD, Neonatology
Oluyinka Olutoye, MD, Pediatric Surgery
Fadel Ruiz, MD, Pulmonary
Cole Burgman, ECLS
Christopher Cassady, MD, Diagnostic Imaging
Milenka Cuevas, MD, Neonatology
Kimberly Dinh, PharmD, Pharmacy
Nikashia Franklin, RN, Nursing
Joseph Garcia-Prats, MD, Neonatology
Jamie Gilley, NP, Neonatology
Amy Hair, MD, Neonatology
Suzanne Iniguez, RT, Respiratory
Gloria Johnson, RN, Nursing
Karen Johnson, MD, Neonatology
Sundeep Keswani, MD, Pediatric Surgery
Kamlesh Kukreja, MD, Diagnostic Imaging
Maria Lantin-Hermosa, MD, Cardiology
Timothy Lee, MD, Pediatric Surgery
Barbara Levy, RN, Nursing
George Mallory, MD, Pulmonary
Barbara McFadden, NP, Neonatology
Amy Mehollin-Ray, MD, Diagnostic Imaging
Erika Moscona, Patient/Family Advisor
Mohan Pammi, MD, Neonatology
Christopher Rhee, MD, Neonatology
Daryl Scott, MD, Genetics
Lois Tracy, NP, Neonatology
Gautham Suresh, MD, Neonatology
Mercy Thundiyil, NP, Neonatology
Nidhy Varghese, MD, Pulmonary
Adam Vogel, MD, Pediatric Surgery
Robert Voigt, MD, Developmental Pediatrics
Allison Walton, RN, Neonatology

EBOC Team
Andrea Jackson, MBA, RN, Research Specialist
Ellis Arjmand, MD, PhD, Associate Medical Director
Charles Macias, MD, MPH, Medical Director

Development Process

This clinical standard was developed using the process outlined in the EBOC Manual. The literature appraisal documents the following steps:

1. Review Preparation
   - PICO questions established
   - Evidence search confirmed with content experts

2. Review of Existing Internal and External Guidelines
   - Guidelines for Acute Care of the Neonate, Baylor College of Medicine Section of Neonatology; Management of CDH Infants, TCH Baylor Physicians; Standardized Postnatal Management of Infants with Congenital Diaphragmatic Hernia in Europe, CDH Euro-Consortium; Surfactant Replacement Therapy for Preterm and Term Neonates with Respiratory Distress, American Academy of Pediatrics; Surfactant Replacement Therapy, American Association of Respiratory Care; NICU Clinical Guidelines Respiratory Problems and Management Congenital Diaphragmatic Hernia, King Edward Memorial Hospital; Management of Congenital Diaphragmatic Hernia: A Systematic Review from the APSA Outcomes and Evidence Based Practice Committee, American Pediatric Surgical Association; CDH Protocol, Cincinnati Children's Hospital; Management of Infants with Congenital Diaphragmatic Hernia from Birth to Surgery, UT Health Science Center at Houston; Congenital Diaphragmatic Hernia Management, Children's Hospital Network; Pediatric Pulmonary Hypertension, American Heart Association and American Thoracic Society; Expert Consensus Statement on the Diagnosis and Treatment of Pediatric Pulmonary Hypertension, The European Paediatric Pulmonary Vascular Disease Network; Pulmonary Hypertension Associated with Acute or Chronic Lung Diseases in Preterm or Term Neonate and Infant, The European Paediatric Pulmonary Vascular Disease Network; General Guidelines for all ECLS Cases, Extracorporeal Life Support Organization; Infection Control and Extracorporeal Life Support, Extracorporeal Life Support Organization; Thorascopic Repair of Congenital Diaphragmatic Hernia in Neonates, National Institute for Health and Clinical Excellence

3. Literature Review of Relevant Evidence
   - Searched: PubMed, Cochrane Library, Google Scholar, Cinahl, Guideline Clearing House

4. Critically Analyze the Evidence
   - 14 meta-analyses, 4 randomized **controlled** trials, and 103 nonrandomized studies

5. Summarize the Evidence
   - Materials used in the development of the guideline, evidence summary, and order sets are maintained in a congenital diaphragmatic hernia evidence-based review manual within EBOC.

Evaluating the Quality of the Evidence

Published clinical guidelines were evaluated for this review using the **AGREE II** criteria. The summary of these guidelines are included in the literature appraisal. AGREE II criteria evaluate Guideline Scope and Purpose, Stakeholder Involvement, Rigor of Development, Clarity and Presentation, Applicability, and Editorial Independence using a 4-point Likert scale. The higher the score, the more comprehensive the guideline.
This clinical standard specifically summarizes the evidence *in support of* or *against* specific interventions and identifies where evidence is *lacking/inconclusive*. The following categories describe how research findings provide support for treatment interventions.
"Evidence Supports" provides clear evidence that the benefits of the intervention exceed harm.
"Evidence Against" provides clear evidence that the intervention is likely to be ineffective or that it is harmful.
"Evidence Lacking/Inconclusive" indicates there is currently insufficient data or inadequate data to support or refute a specific intervention.

The **GRADE** criteria were utilized to evaluate the body of evidence used to make practice recommendations. The table below defines how the quality of the evidence is rated and how a strong versus weak recommendation is established. The literature appraisal reflects the critical points of evidence.

Recommendation	
STRONG	Desirable effects clearly outweigh undesirable effects or vice versa
WEAK	Desirable effects closely balanced with undesirable effects
Quality	**Type of Evidence**
High	Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies
Moderate	Evidence from RCTs with important limitations (e.g., inconsistent results, methodological flaws, indirect evidence, or imprecise results) or unusually strong evidence from unbiased observational studies
Low	Evidence for at least 1 critical outcome from observational studies, RCTs with serious flaws or indirect evidence
Very Low	Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence

Recommendations

Practice recommendations were directed by the existing evidence and consensus amongst the content experts. Patient and family preferences were included when possible. The Content Expert Team and EBOC team remain aware of the controversies in the diagnosis/management of congenital diaphragmatic hernia in children. When evidence is lacking, options in care are provided in the clinical standard and the accompanying order sets (if applicable).

Approval Process

Clinical standards are reviewed and approved by hospital committees as deemed appropriate for its intended use. Clinical standards are reviewed as necessary within EBOC at Texas Children's Hospital. Content Expert Teams are involved with every review and update.

Disclaimer

Practice recommendations are based upon the evidence available at the time the guideline was developed. Clinical standards (guidelines, summaries, or pathways) do not set out the standard of care, and are not intended to be used to dictate a course of care. Each physician/practitioner must use his or her independent judgment in the management of any specific patient and is responsible, in consultation with the patient and/or the patient family, to make the ultimate judgment regarding care.

Appendix

The observed-to-expected total lung volume (O/E TFLV) and percent liver herniation (%LH) has been used for prenatal risk stratification in patients with congenital diaphragmatic hernia [(20)]. Below are the risk stratification categories utilized at Texas Children's Hospital.

Severity	O/E TFLV	LH%
Mild	>32%	<21%
Moderate	≥32%	>21%
Moderate	<32%	≤21%
Severe	<32%	>21%

Related Documents

Congenital Diaphragmatic Hernia Literature Appraisal Document